[CASE REPORT AND LITERATURE REVIEW]

Atypical Presentation of Cutaneous Lupus Mucinosis

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ABSTRACT

Objective: To describe the epidemiological, clinical, and serological features of cutaneous lupus mucinosis and its relationship to systemic lupus erythematosus as well as elucidate the histopathological features of cutaneous lupus mucinosis and describe how these features differ from tumid lupus erythematosus. **Design:** Case review and assessment of the literature. **Setting:** University academic setting. **Participant:** One patient. **Results:** The authors report a case of antinuclear antibody negative cutaneous lupus mucinosis in a patient without systemic involvement who responded to hydroxychloroquine and intralesional triamcinolone. A review of the literature found 30 reported cases of cutaneous lupus mucinosis, three of which were antinuclear antibody negative and the majority had systemic lupus erythematosus. The most common therapy reported is systemic corticosteroids; however, the patient described in this case had significant improvement of the lesions with only intralesional steroid injections and hydroxychloroquine. Considering the proportion of patients with cutaneous lupus mucinosis who progress to systemic lupus is uncertain, the authors suggest following these patients closely for evidence of multisystem disease. **Conclusion:** The authors report a case of antinuclear antibody-negative cutaneous lupus mucinosis in a patient without systemic lupus erythematosus who responded to hydroxychloroquine and intralesional triamcinolone. Given the rarity of this condition and reported association with systemic lupus erythematosus, it is important to follow these patients clinically for any signs or symptoms of systemic involvement. (*J Clin Aesthet Dermatol.* 2013;6(4):37–40.)

A 38-year-old Vietnamese man presented with a two-year history of several painful, slightly scaly, erythematous nodules first appearing on the chest and later on the bilateral posterior arms. The lesions had significantly increased in size and became painful in the prior three months causing the patient to seek treatment. His past medical history was unremarkable and he denied any systemic signs or symptoms including fever, oral ulceration, photosensitivity, or arthralgia. He reported no family history of lupus erythematosus or other connective tissue disease. Physical examination revealed multiple, tender, firm, erythematous nodules with fine scale on the anterior chest and posterior arms (Figure 1). No ocular or oral ulcerations were noted. Laboratory evaluation showed a normal complete blood count, basic metabolic panel, serum creatinine, liver function tests, urinalysis, thyroid stimulating hormone level, antinuclear antibody (ANA), and rheumatoid factor. G6PD testing was within normal limits. An incisional biopsy specimen of the left arm showed a dense superficial and deep perivascular and periadnexal lymphocytic infiltrate in the dermis with overlying parakeratosis and focal vacuolar alteration of the dermal-epidermal junction (Figures 2A and 2B). The entire dermis and subcutaneous fat was completely replaced with mucinous material (Figures 3A and 3B). There was no evidence of vasculitis. The patient was treated with hydroxychloroquine 200mg twice daily and intralesional triamcinolone acetonide 10mg/cc. At four-week follow up, there was significant flattening of the lesions and decreased erythema and near resolution of tenderness. The patient did not develop any new lesions in the interim. Originally described by Gold[1] in a patient with systemic lupus erythematosus who presented with white, lumpy lesions on the neck and back, cutaneous lupus mucinosis (CLM) is a rare, distinct subset of chronic cutaneous lupus erythematosus. Recent reports in the

DISCLOSURE: The authors report no relevant conflicts of interest.
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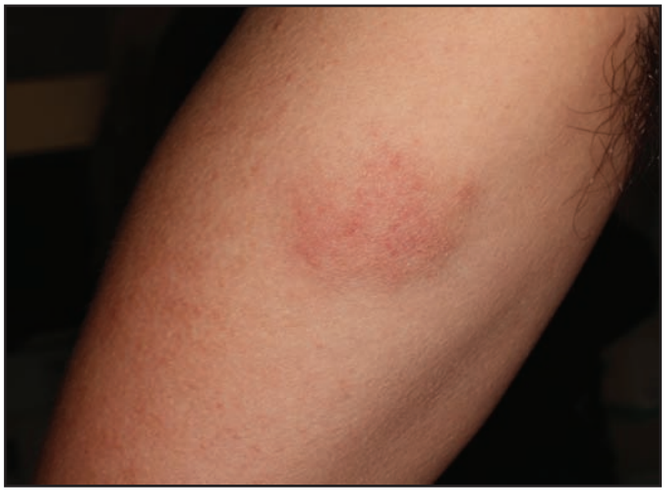
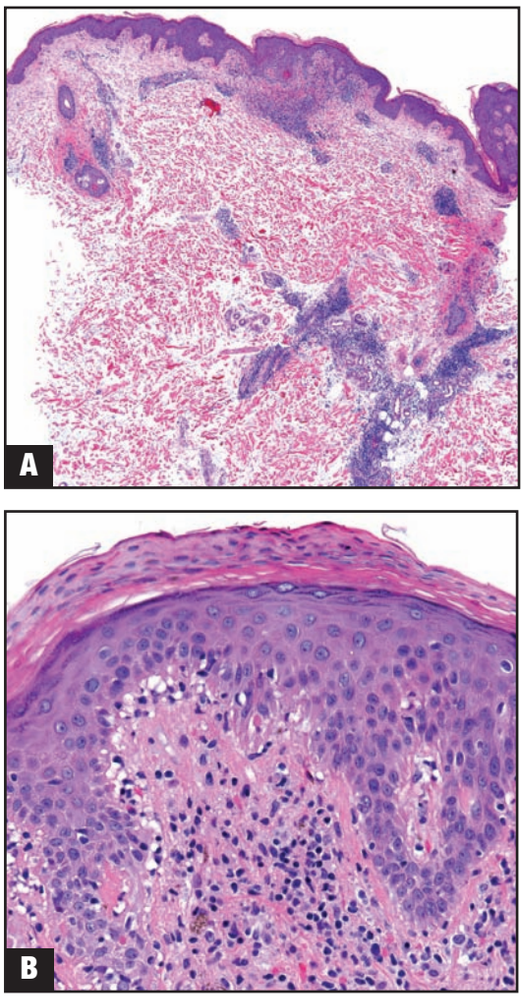


Figure 1. Well-circumscribed erythematous tumor with subtle scale on the triceps

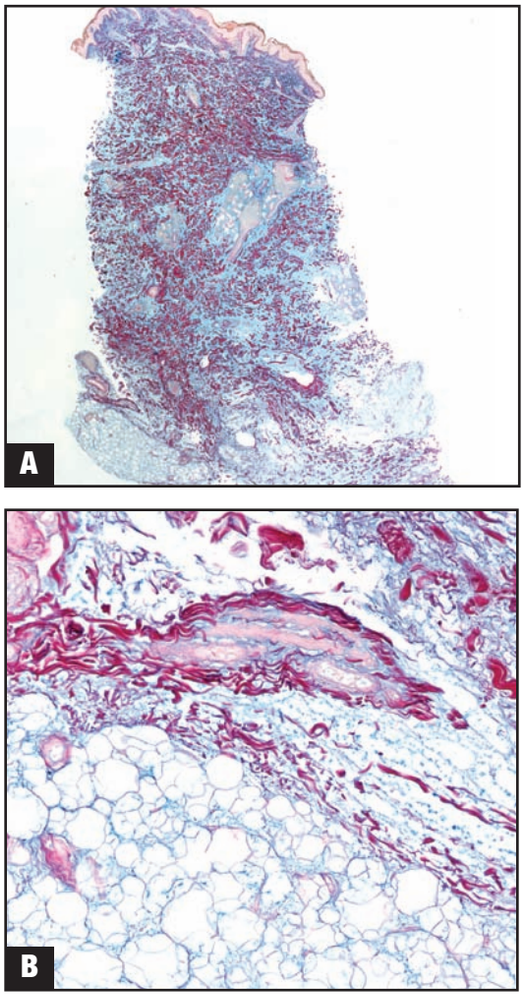


Figures 2A and 2B. Scanning view (A, 40x) showing dense superficial and deep perivascular and periadnexal mononuclear cell infiltrates with focal parakeratosis and subtle vacuolar interface dermatitis. High-power (B, 400x) magnification demonstrating subtle vacuolar interface dermatitis with overlying parakeratosis and pigment incontinence.

literature have sought to elucidate the specific clinical and histopathological characteristics of CLM.

The authors reviewed the 30 known cases of CLM reported in the English language by PubMed search and derived demographic, clinical, and histopathological data to help define this clinical entity. In this cohort, 47 percent were male and 53 percent were female. Interestingly, in Caucasian patients, there was a female preponderance while in Asian patients, males predominated. The significance of this finding is unknown. The average age of presentation is 37.6 years with two-thirds of cases presenting in the fourth and fifth decades of life. There was no significant difference in the age of onset when comparing males to females. The range of presentation of CLM is highly variable; the most common presentation being multiple skin-colored nodules varying in size. Moreover, there is a report of one case presenting with profound mucinosis in tumorous masses[1] and another with just a single nodule on the shoulder.[2] In terms of distribution, 87 percent of patients with CLM had lesions on multiple body areas. The most common locations are the back and upper arms, but lesions may be seen on the face, neck, chest, hands, breasts, and the lower extremities. CLM has a very strong association with systemic lupus erythematosus (SLE). Based on this review, 77 percent of patients with CLM had met 4 of the 11 criteria defined by the American College of Rheumatology for SLE prior to presentation.[1–4,8–23] However, not all of these patients had been given the diagnosis of SLE; some patients with CLM had, by history, demonstrated signs and/or symptoms of SLE or discoid lupus erythematosus (DLE) months before actual presentation, while other cases had nonspecific symptoms (arthralgia, proteinuria, and malar rash) that were postulated to be early features of systemic disease. Only one documented case of CLM has been asymptomatic and without evidence of systemic disease.[4] In the literature, ANA positivity in CLM has been observed in 25 of 30 cases (83%), while 3 of 30 were ANA negative and 2 of 30 were not reported. The case described herein is atypical in that it occurred in a patient with no prior history of lupus erythematosus and is ANA negative. Thus, considering that most reports suggest a relationship between SLE and CLM, it is imperative to follow these patients clinically to ascertain new signs or symptoms consistent with SLE, although the risk of progression to systemic disease has not been firmly established. Although the etiopathogenesis of CLM is unknown, numerous hypotheses have been reported, including an abnormal immune response linked to human leukocyte antigens (HLA) B8 and DR3 haplotypes, increased glycosaminoglycan production by dermal fibroblasts stimulated by an unidentified cytokine,[5] an increase in mucinoses-related cytokines by male sex hormones,[6] or suppression of mucin degradation.[7] The diagnosis of CLM can be supported on the basis of the unique histological features. Prominent findings include a dense perivascular and periadnexal lymphocytic infiltrate with striking pandermal and subcutaneous masses of mucinous material

with or without interface dermatitis. Recent reports have also documented vasculitis in CLM.[12] Histopathologically, CLM should be differentiated from tumid lupus erythematosus (TLE). TLE typically lacks interface changes, although there are reports of focal interface changes at the dermal-follicular and the dermal-epidermal junction.[23] In CLM, interface changes tend to be more common and have been documented in 13 of 30 patients to date.[1–4,8–23] Moreover, only 7 of 30 cases have not demonstrated interface dermatitis, while in 10 of 30, the presence or absence of interface dermatitis was not documented.[1–4,8–23] The massive amounts of mucin spanning the entire dermis observed in CLM differs from TLE, which is present in much lesser quantities and has a more interstitial distribution. Numerous therapeutic approaches have been used to treat CLM, including indomethacin, mepacrine, chloroquine, topical steroids, intralesional triamcinolone (5mg/cc), prednisolone, prednisone, sulphonamide, salicylates, adrenocorticotropic hormone (ACTH), bismuth, gold injections, hyaluronidase, and hydroxychloroquine.[1–4,8–23] The response to systemic corticosteroids (up to 60mg of prednisone daily) has been observed in 13 reported cases, with significant improvement in the cutaneous lesions in all patients.[1–4,8–23] In 11 cases, the use of antimalarials showed efficacy in six patients, while four showed no response and one showed a partial and temporary response. Only one case in the literature reported intralesional injection with triamcinolone, which showed some efficacy. In the patient described herein, a reduction in the erythema and size was noted after one month of hydroxychloroquine 200mg twice daily and intralesional triamcinolone 10mg/cc. This case demonstrates the efficacy of low-dose intralesional triamcinolone combined with hydroxychloroquine for the treatment of CLM while avoiding the use, and serious side effects of, systemic corticosteroids. In conclusion, the authors report a case of ANA-negative cutaneous lupus mucinosis in a patient without SLE who responded to hydroxy-chloroquine and intralesional triamcinolone. Given the rarity of this condition and reported association with SLE, it is important to follow these patients clinically for any signs or symptoms of systemic involvement.



Figures 3A and 3B. Colloidal iron stain demonstrating pandermal and subcutaneous mucopolysaccharide deposition (A, 20x) and alteration of subcutaneous lipocytes with abundant mucin (B, 200x).

REFERENCES

1. Gold SC. An unusual papular eruption associated with lupus erythematosus. *Br J Dermatol.* 1954;66(12):429–433.
2. Maruyama M, Miyauchi S, Hashimoto K. Massive cutaneous mucinosis associated with systemic lupus erythematosus. *Br J Dermatol.* 1997;137(3):450–453.
3. Weigand DA, Burgdorf WH, Gregg LJ. Dermal mucinosis in discoid lupus erythematosus. Report of two cases. *Arch Dermatol.* 1981;117(11):735–738.
4. Obermoser G, Sontheimer RD, Zelger B. Overview of common, rare and atypical manifestations of cutaneous lupus erythematosus and histopathological correlates. *Lupus.* 2010;19(9):1050–1070.
5. Pandya AG, Sontheimer RD, Gockerell GJ, et al. Papulonodular mucinosis associated with systemic lupus erythematosus: possible mechanisms of increased glycosaminoglycan accumulation. *J Am Acad Dermatol.* 1995;32(2):199–205.
6. Kim Yi, Kim JH, Zelner DJ, et al. Transforming growth factor-beta 1 is a mediator of androgen regulated growth arrest in an androgen-responsive prostatic cancer cell line, LNGaP. *Endocrinology.* 1996;137(3):991–999.
7. Noguchi T, Igarashi M, Saitoh S. A case of lichen myxoedematosis associated with SLE. *Rinsho Derma.* 1980;22:1223 [in Japanese].
8. Rajan N, Wahie S, Seukeran D. Papulonodular eruption, proteinuria, and peripheral arthritis. Papulonodular dermal mucinosis (PDM) in systemic lupus erythematosus. *Arch Dermatol.* 2010;146(7):789–794.
9. Terao H, Moroi Y, Urabe K, et al. A case of nodular cutaneous lupus mucinosis. *J Dermatol.* 2003;30(4):341–343.
10. Sonntag M, Lehmann P, Megahed M, Ruzicka T, Kuhn A. Papulonodular mucinosis associated with subacute cutaneous lupus erythematosus. *Dermatology.* 2003;206(4):326–329.
11. Rongioletti F, Rebora A. Cutaneous mucinoses: microscopic

criteria for diagnosis. *Am J Dermatopathol.* 2001;23(3): 257–267.
12. Kanda N, Tsuchida T, Watanabe T, Tamaki K. Cutaneous lupus mucinosis: a review of our cases and the possible pathogenesis. *J Cutan Pathol.* 1997;24(9):553–558.
13. Kano Y, Sagawa Y, Yagita A, Nagashima M. Nodular cutaneous lupus mucinosis: report of a case and review of previously reported cases. *Cutis.* 1996;57(6):441–444.
14. Jackson BA, Norum RA, Lowe L, Fivenson DP. Papulonodular dermal mucinosis in systemic lupus erythematosus. *J Rheumatol.* 1994;21(5):940–941.
15. Choi EH, Hann SK, Chung KY, Park YK. Papulonodular cutaneous mucinosis associated with systemic lupus erythematosus. *Int J Dermatol.* 1992;31(9):649–652.
16. Lowe L, Rapini RP, Golitz LE, Johnson TM. Papulonodular dermal mucinosis in lupus erythematosus. *J Am Acad Dermatol.* 1992;27(2 Pt 2):312–315.
17. Rongioletti F, Parodi A, Rebora A. Papular and nodular mucinosis as a sign of lupus erythematosus. *Dermatologica.* 1990;180(4):221–223.
18. Rongioletti F, Rebora A. Papular and nodular mucinosis associated with systemic lupus erythematosus. *Br J Dermatol.* 1986;115(5):631–636.
19. Fowler JF Jr, Callen JP. Cutaneous mucinosis associated with lupus erythematosus. *J Rheumatol.* 1984;11(3):380–383.
20. Lamberts RJ, Lynch PJ. Nodular cutaneous mucinosis associated with lupus erythematosus. *Cutis.* 1981;28(3):294–295,297–299.
21. Kennedy C, Gold SC, Moss R. Systemic lupus erythematosus with an unusual popular eruption. *Br J Dermatol.* 1979;101(Suppl 17):51–53.
22. Gammon WR, Caro I, Long JC, Wheeler CE Jr. Secondary cutaneous mucinosis with systemic lupus erythematosus. A case presentation and review of the literature. *Arch Dermatol.* 1978;114(3):432–435.
23. Alexiades-Armenakas MR, Baldassano M, Bince B, et al. Tumid lupus erythematosus: criteria for classification with immunohistochemical analysis. *Arthritis Rheum.* 2003;49(4):494–500.